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Keywords: Myopathies; Facioscapulohumeral dystrophy; Exercise therapy; Maximal strength; Fatigue resistance; Aerobic power; Quality of life; Fatigue and functional tests

Background.– It is now accepted that physical activity is not deleterious in myopathies, including muscular dystrophies. Facioscapulohumeral dystrophy (FSHD) is characterized by a progressive decline of muscular function with time, associated with a significant loss of autonomy and chronic fatigue. Only a few controlled and randomized studies have been conducted on exercise therapy and mainly performed on short supervised periods. This study aimed to evaluate the effects of a 24-weeks of a home-based and coached exercise program on motor function in FSHD patients.
Methods.– Sixteen FSHD patients randomized (trained/controls) were recruited in the Rhône-Alpes Reference Centre for Rare Neuromuscular Diseases consultations. The training group realized three weekly sessions on cycle. Multi-factorial evaluations, before, after and over the training period were performed in both groups: maximal strength, fatigue resistance, aerobic power, questionnaires of quality of life and functional tests. Biopsies from vastus lateralis muscles were done (beginning and end of the program). Program started under a coach supervision and was monitored by heart rate recordings.
Results.– Patients performed this program of exercise therapy with a great assiduity and an excellent tolerance. We report multi-factorial benefits of this home-based training program and its influence on the quality of life of trained patients.

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Evaluation of muscle oxygenation in patients with facioscapulohumeral muscular dystrophy

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Keywords: Neuromuscular disease; Muscle oxygenation; Near-infrared spectroscopy; Physical exercise

Objective.– To determine changes in muscle oxygenation during effort in patients with facioscapulohumeral muscular dystrophy (FSH).
Methods.– Eight FSH patients and 15 age-matched controls performed isokinetic constant-load knee extension exercises at 20% of their extensors' peak torque (i.e. the same relative load) for up to 4 min. All exercises consisted in rhythmic, voluntary, isokinetic, concentric contractions of the quadriceps at 90°/s, whereas the return was performed passively at the same speed. Muscle oxygenation in the vastus lateralis was evaluated noninvasively using near-infrared spectroscopy (NIRS). During exercise, deoxygenated hemoglobin (HHB) and blood volume were significantly lower in the FSH patients. The initial muscle deoxygenation and functional impairment (walking endurance) were correlated with the peak torque.
Discussion.– The findings in this study suggest that FSH subjects present an impairment in their capacity to deliver or to use oxygen and would be the consequences of the deconditioning syndrome.
Further reading
Olsen D, et al. Aerobic training improves exercise performance in facioscapulohumeral muscular dystrophy. Neurology 2005;64:1064–1066.

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Rasch analysis of the motor function measure in patients with congenital muscle dystrophy and congenital myopathy

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Keywords: Dystrophie musculaire congénitale; Myopathie congénitale; Évaluation de la fonction motrice; Mesure des résultats; Analyse Rasch

Objectives.– Valid outcome measures are necessary to monitor the treatment effects in patients with congenital disorders of muscle.
Methods.– In 19 departments in France, Belgium, and USA, 289 patients aged 5- to 77-years-old were enrolled. A Rasch analysis examined the robustness of the motor function measure across the disease spectrum. The three domains (standing position and transfers, axial and proximal motor function, and distal motor function) were examined using RUMM 2030 software with a partial credit model.
Results.– The original 32-item MFM did not fit the Rasch model expectations enough in neither of its domains. Switching from a four- to a three-category response-scale in 18 items restored response order in 16. Various additional checks suggested the removal of seven items. The resulting 25-item MFM demonstrated a good fit to the Rasch model. Domain 1 was well-targeted to the whole severity spectrum whereas domains 2 and 3 were better targeted to severe cases. The reliability coefficients MFM-25 suggested sufficient ability for each summed score to distinguish between patient groups (0.9, 0.8, and 0.7 for domains 1, 2, and 3, respectively).
Discussion.– The Rasch-scaled MFM-25 can be assumed to be a linear scale in each of its three domains.

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